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Modern Concepts of Cardiovascular Disease

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CONGENITAL HEART DISEASE IN CHILDHOOD WITH SPECIAL REFERENCE TO PROGNOSIS

Accurate information as to morbidity and mortality resulting from congenital cardiac anomalies is difficult to obtain. It is a natural tendency to report the striking and the unusual. Compilations of reported cases therefore probably overemphasize complicated abnormalities and deaths due to dramatic causes such as bacterial endocarditis, cerebral abscess or hemorrhage, rupture of the aorta, etc.

Statistics based on patients within a hospital will be influenced by the fact that these individuals represent a group chosen because of the existence of symptoms sufficient in degree to require hospital treatment. Impressions derived from an experience with school children on the other hand will be unduly optimistic, since the highest death rate in childhood seems to occur within the first two years of life. Ideally, the outcome of cardiac anomalies could best be determined by the continuous observation of unselected groups of newborn infants — a task phenomenal in extent when we take into consideration the fact that the incidence of such anomalies among the newborn is probably well under one per cent.

That the mortality rate among groups of children with congenital cardiac defects will be influenced greatly by the selection of patients is emphasized by a follow-up study made in 1938 of over two hundred children with a presumptive diagnosis of cardiac anomaly seen at the Children's Hospital of Philadelphia during the years 1922 to 1936. Diagnosis was based on the existence of murmurs best heard at sites not characteristic of the acquired valvular lesions, aided by the roentgenologic finding of cardiac enlargement or change in contour; by abnormalities noted in the electrocardiogram; and in some of the cases by the presence of a precordial thrill, cyanosis or clubbing of the fingers. Since in the last analysis the anatomic diagnosis can be established only at the autopsy table, for the purpose of this study no attempt was made to estimate the outcome of the individual anatomic lesions.

One-half of the children had never been cyanotic; approximately one-third had presented persistent cyanosis; the remainder gave a history of transient cyanosis in early life or had manifested cyanosis for a relatively brief period preceding death.

Forty-seven per cent of the group had died, deaths being equally divided between boys and girls.

Seventy-three per cent of these deaths had occurred within the first year of life and seventy-eight per cent within the first two years.

Eighty-four per cent of all deaths had occurred among patients first seen on the wards. The majority of these children had been admitted with symptoms of serious illness. Only sixteen per cent of the deaths had occurred among cases originally admitted to the cardiac clinic, where children were kept under observation because of the existence of a cardiac lesion, regardless of the existence of symptoms. In many of these children followed in the clinic, the lesion had been discovered accidentally during the course of a routine physical examination.

Of the 72 children who had been admitted to the wards as babies, ninety-one per cent were dead in 1938. The outcome for babies with cardiac anomaly and illness severe enough to necessitate hospital admission would seem to be gloomy. Of the 42 infants who had originally come under the observation of the cardiac clinic, one-third had died. The death rate would therefore seem to be high even in a group of infants assembled only because of the existence of a cardiac defect. Among the 94 children who had reached school age the incidence of death was eleven per cent.

The death rate of cyanotic infants during the first year of life was twice that of non-cyanotic and rose progressively as the children gradually succumbed to infection and circulatory failure. After the second year of life there was a marked drop in deaths among non-cyanotic individuals. The ratio of cyanotic to non-cyanotic children on the wards was 1:2; in the clinic, 1:4; among children of school age, 1:6. Only five per cent of the non-cyanotic school children had died as compared with eighty per cent of the cyanotic individuals of school age.

Cause of death. In slightly over one-half the cases the final illness was of infectious origin. Pneumonia headed the list of infections, causing thirty-nine per cent of the deaths. Bacterial endocarditis was the terminal event in only two instances. In one-third of

the cases circulatory inadequacy was a prominent feature.

Of the 38 children who came to necropsy, fifty-seven per cent of the cyanotic group had died of infection and forty-three per cent of the cardiac defect itself. In the acyanotic and delayed cyanotic groups, eighty-eight per cent had died of infection and one child (six per cent) of circulatory failure. In addition, a nine-year-old girl with aortic stenosis had died suddenly following a blow over the precordium received in a childish quarrel.

Clinical Aspects. Infants with congenital cardiac defects tend to be slow to gain weight and delayed in growth. The marasmus of the infants admitted to the wards undoubtedly was a factor in the high mortality rate. In later life these children may remain dwarfed and of fragile build. In some individuals, however, a spurt of growth occurs as puberty is reached.

Mental deficiency is a not infrequent occurrence. Fourteen per cent of the present group were mentally retarded, of whom over half were Mongolian idiots or microcephalic.

In infancy and early childhood increased susceptibility to infections, especially respiratory, is common. Pneumonia, though a frequent cause of death, is not invariably fatal. It is known that 23 of our children (one cyanotic) survived attacks of pneumonia. Several individuals had suffered repeated illnesses in early life associated with signs of pulmonary involvement. Since there was a high incidence of lipoid pneumonia among the patients examined postmortem it is possible that some of these children with long-continued pneumonia in early childhood had suffered from lipoid pneumonia. No death occurred from any of the infectious diseases common in childhood, including pertussis in 11 cases. Eight children had suffered from rheumatic infection of mild degree.

Operative procedures were well borne, including 33 tonsillectomies and adenoidectomies (two in cyanotic individuals); one splenectomy, one mastoidectomy, one herniorrhaphy, one appendectomy, one open reduction for subluxation of the elbow. Recently a patent ductus arteriosus was ligated successfully in one patient.

There are many instances of longevity among non-cyanotic individuals with congenital cardiac defects, but on the whole the prognosis is poor for the cyanotic individual. In the present series, of 16 children with persistent cyanosis who reached the age of five years, 10 were leading lives of greatly restricted activity. Of the 7 who reached the age of ten years, 4 were cardiac cripples. Four children lived past their fifteenth birthday. One has since died; two have always been incapable of much exertion.

In an occasional instance, however, the outlook is by no means hopeless. Dr. Paul White has reported the case of the talented musician with tetralogy of Fallot who died in his sixtieth year. There are various such reports of cyanotic individuals who have lived long and useful lives, sometimes even of an arduous nature. Cyanotic women have given birth to normal children.

The oldest cyanotic individual in the present series is a boy, 23 years of age, intelligent and with many interests, employed as a laboratory technician. Eight years ago this boy's erythrocyte count was 9,000,000 and his hemoglobin was 150 per cent, which would seem to controvert the statement quoted by Abbott that a count above 6,000,000 is always of grave prognostic import. In the group of living cyanotic children in this series the average hemoglobin was 118 per cent, the average erythrocyte count 7,390,000, the maximum hemoglobin 165 per cent and the maximum erythrocyte count 11,180,000. Of the cyanotic individuals who had died, the average erythrocyte count was 5,380,000, the average hemoglobin 90 per cent, the maximum erythrocyte count 7,820,000 and the maximum hemoglobin 128 per cent. Presumably death had occurred before there had been an opportunity to attain maximum polycythemia.

Since it is possible for an occasional cyanotic individual to attain a self-supporting adult life, it is unfair to assume a hopeless prognosis in any child who has survived infancy. Cyanotic children should not be deprived of schooling.

Whether cyanosis is present or absent, there is no indication for exercise restriction beyond the limiting capacity of the heart itself. Children with congenital lesions are more fortunate than their rheumatic brothers and sisters in that they do not have the additional handicap of continued systemic and myocardial infection. In spite of startling murmurs and cardiac enlargement, the functional capacity is often normal. Of the 115 living patients in the present series (twenty-five per cent of whom were over 15 years of age; twenty-seven per cent between the age of 10 and 15; forty-eight per cent below 10 and over 2); 89 were able to carry on a normal existence; 15 were slightly restricted in their activity; and 10 — all cyanotic — were greatly limited in their ability to undergo exertion.

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